Case Reports

Sudden Unilateral Vision Loss

Arising from Calcified Amorphous Tumor of the Left Ventricle

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Calcified amorphous tumor of the heart is a very rare non-neoplastic intracavitary mass. The clinical presentation is similar to that of other cardiac masses. The precise cause and best approach to treatment remain unclear. We describe a case of cardiac calcified amorphous tumor presenting with refractory unilateral vision loss that was successfully treated by surgical excision. To our knowledge, this is only the 2nd reported case of retinal arterial embolism due to cardiac calcified amorphous tumor in the English-language literature.
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Calcified amorphous tumor (CAT) of the heart is a very rare non-neoplastic cardiac intracavitary mass. It was first described as a distinct pathologic entity by Reynolds and colleagues in 1997.[1] Since then, very few cases of cardiac CAT have been reported in the English-language medical literature[2-19] (Table I). The clinical presentation is similar to that of other cardiac masses. The precise cause of the mass and the best means of treatment remain unclear.[12]

In the present report, we describe a case in which cardiac CAT presented with refractory unilateral vision loss, which was successfully treated by surgical excision.

Case Report

A 54-year-old woman presented to her local physician with unilateral vision loss (left eye), which had occurred suddenly 27 days before. The patient was referred to our hospital. Left central retinal arterial occlusion was detected upon fundus examination (Fig. 1A), and we set about to identify the source of the embolus. The results of her physical examination were otherwise normal. She had a history of hypothyroidism and was under treatment with oral levothyroxine. Laboratory tests, including tests for parathyroid hormone, thrombotic, and autoimmune irregularities, yielded results within normal limits. The electrocardiogram was also unremarkable. A plain chest film showed a localized dense calcified mass within the cardiac silhouette. Transthoracic echocardiography (TTE) then revealed a pedunculated mobile hyperechoic calcified mass 3.8 × 2.5 cm in size, which arose from the septoapical and anteroapical region of the left ventricle (Figs. 1B–D). No right-to-left shunt was detected by TTE. No laboratory data suggested infective endocarditis. Anticoagulative therapy was immediately started. A computed tomographic scan confirmed the presence of a heavily calcified, irregularly shaped mass 3.5 × 2.6 cm in size, which arose from the region of the left ventricular endomyocardium as indicated by TTE (Figs. 1E, 1F, and 2A). Duplex ultrasonographic imaging of the carotid and vertebral arteries yielded unremarkable results. Conventional coronary angiography showed normal coronary arteries. Brain magnetic resonance imaging also showed nothing of note.

Our patient underwent cardiac exploration and removal of the mass to prevent further systemic embolization. A median sternotomy was performed. The cannulations for cardiopulmonary bypass were done via the ascending aorta and the right atrial appendage. After cross-clamping the ascending aorta, we arrested the heart with cold-blood cardioplegic solution administered antegrade under mild hypothermia. An anteroapical left ventriculotomy was then performed. Intraoperatively, we found a calcified mass adjacent to the septoapical and anteroapical region of the left ventricle. The large, rough, fragile mass was excised in toto (but as multiple pieces) through a left ventriculotomy (Fig. 2B). Perioperative transesophageal echocardiography detected no

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TABLE I. Clinicopathologic Findings of Published Cases of Cardiac Calcified Amorphous Tumor

Report	Case No.	Age (yr); Sex; Clinical Presentation	Diagnostic Procedure(s)	Tumor Site	Treatment
Reynolds C, et al.[1] (1997)	1	16; M; exercise-associated headache, near-syncope, and exercise intolerance	TTE and MRI	Left atrium	Surgical excision
	2	30; M; near-syncope, chest pain, and palpitations	Not known	LV	Surgical excision
	3	33; F; shortness of breath	TTE and MRI	RV	Surgical excision
	4	34; F; vertigo and orthopnea	TTE and MRI	RV	Surgical excision
	5	48; F; CVA	TTE	Mitral valve	Surgical excision
	6	60; F; CVA and retinal emboli	TTE and TEE	LV	Surgical excision
	7	65; M; shortness of breath	Not known	RV	No surgery
	8	67; F; syncope	Not known	Right atrium	Surgical excision
	9	67; M; syncope	TTE	LV	Surgical excision
	10	73; F; dizziness and dyspnea on exertion	TTE, TEE, and cardiac catheterization	Right atrium and SVC	Surgical excision
	11	75; F; funny sensation in chest	Fluoroscopy and TTE	LV	Surgical excision
Chaowalit N, et al.[2] (2005)	1	20; F; pulmonary embolism	TTE, CT, MRI, and histopathologic examination	RV (apex)	Surgical excision
Lewin M, et al.[3] (2006)	1	60; F; syncope	TTE, MRI, cardiac catheterization, and histopathologic examination	RV (ventricular lateral wall and annulus of the TV)	Surgical radical excision and TVR
Fealey ME, et al.[4] (2007)	1	23; F; pulmonary embolization	CT, TTE, TEE, and histopathologic examination	RV (apex)	Surgical excision
Khulbey S, et al.[5] (2008)	1	26; M; prolonged fever and constitutional symptoms	TTE and histopathologic examination	Right atrial free wall	Surgical excision
Ho HH, et al.[6] (2008)	1	44; M; progressive shortness of breath on exertion	Chest radiography, TEE, CT, cardiac catheterization, and histopathologic examination	LV (diffuse infiltration of the ventricular myocardium, papillary muscles, and mitral chordal apparatus)	Referred for possible heart transplantation
Gutierrez-Barrios A, et al.[7] (2008)	1	35; M; septic shock	TTE, TEE, and histopathologic examination	Along right atrial wall and eustachian valve of IVC	Surgical excision
Flynn A and Mukherjee G[8] (2009)	1	"Young"; M; syncope	TTE, MRI, and histopathologic examination	RV (chordae tendineae of TV)	Surgical excision and pulmonary thrombectomy
Habib A, et al.[9] (2010)	1	58; F; recurrent ventricular tachycardia	Chest radiography, TTE, CT, electrophysiologic study, and ICE	LV (endocardium, papillary muscles, and mitral annulus)	Combination antiarrhythmic therapy

TABLE I, continued. Clinicopathologic Findings of Published Cases of Cardiac Calcified Amorphous Tumor

Report	Case No.	Age (yr); Sex; Clinical Presentation	Diagnostic Procedure(s)	Tumor Site	Treatment
Vaideeswar P, et al.[10] (2010)	1	56; M; progressive shortness of breath, blurring of vision	TTE, MRI, and histopathologic examination	Right atrium	Surgical excision and pulmonary thrombectomy
	2	35; M; exertional dyspnea and dizziness on walking	TTE, CT, MRI, and histopathologic examination	Right atrium near inferior cavoatrial junction	Surgical excision and pulmonary thrombectomy
Gupta R, et al.[11] (2010)	1	40; F; progressive dyspnea, fatigue, and cough	TTE and histopathologic examination	Right atrial wall and septum	Surgical excision
Kubota H, et al.[12] (2010)	1	64; F; incidentally	TEE, CT, and histopathologic examination	LV (mitral valve anterior annulus)	Surgical excision
	2	44; M; incidentally	TTE, CT, and histopathologic examination	LV (anterior papillary muscle of mitral valve)	Surgical excision
Ananthakrishna R, et al.[13] (2011)	1	45; F; breathlessness	Chest radiography, TTE, CT, coronary angiography, and histopathologic examination	LV posterior wall	Surgical excision and MVR
Greaney L, et al.[14] (2011)	1	69; F; left-sided heart failure and stroke	TTE, coronary angiography, and histopathologic examination	LV (base of mitral valve)	Surgical excision
Vlasseros I, et al.[15] (2011)	1	65; F; visual disturbances	TTE, TEE, and histopathologic examination	LV (mitral valve)	Surgical excision and MVR
Lin YC, et al.[16] (2011)	1	74; F; incidentally	Chest radiography, TTE, MRI, coronary angiography, and histopathologic examination	Left atrium	Surgical excision
Sousa JS, et al.[17] (2011)	1	17; M; cardiomegaly	Chest radiography, TTE, TEE, and histopathologic examination	TV	Surgical excision and TV annuloplasty
Fujiwara M, et al.[18] (2012)	1	58; M; incidentally	TTE, TEE, and histopathologic examination	Left atrium and LV (mitral annulus) (2 masses)	Surgical excision
	2	65; M; incidentally	TTE, TEE, and histopathologic examination	LV (mitral annulus)	Surgical excision
Nishigawa K, et al.[19] (2012)	1	78; F; incidentally	TTE, TEE, and histopathologic examination	Left atrium (mitral annulus)	Surgical excision
Current case (2013)	1	54; F; retinal arterial occlusion	Chest radiography, CT, TTE, and histopathologic examination	LV (septoapical and anteroapical regions)	Surgical excision

CT = computed tomography; CVA = cerebrovascular accident; F = female; ICE = intracardiac echocardiography; IVC = inferior vena cava; LV = left ventricle; M = male; MRI = magnetic resonance imaging; MVR = mitral valve replacement; RV = right ventricle; SVC = superior vena cava; TEE = transesophageal echocardiography; TTE = transthoracic echocardiography; TV = tricuspid valve; TVR = tricuspid valve replacement

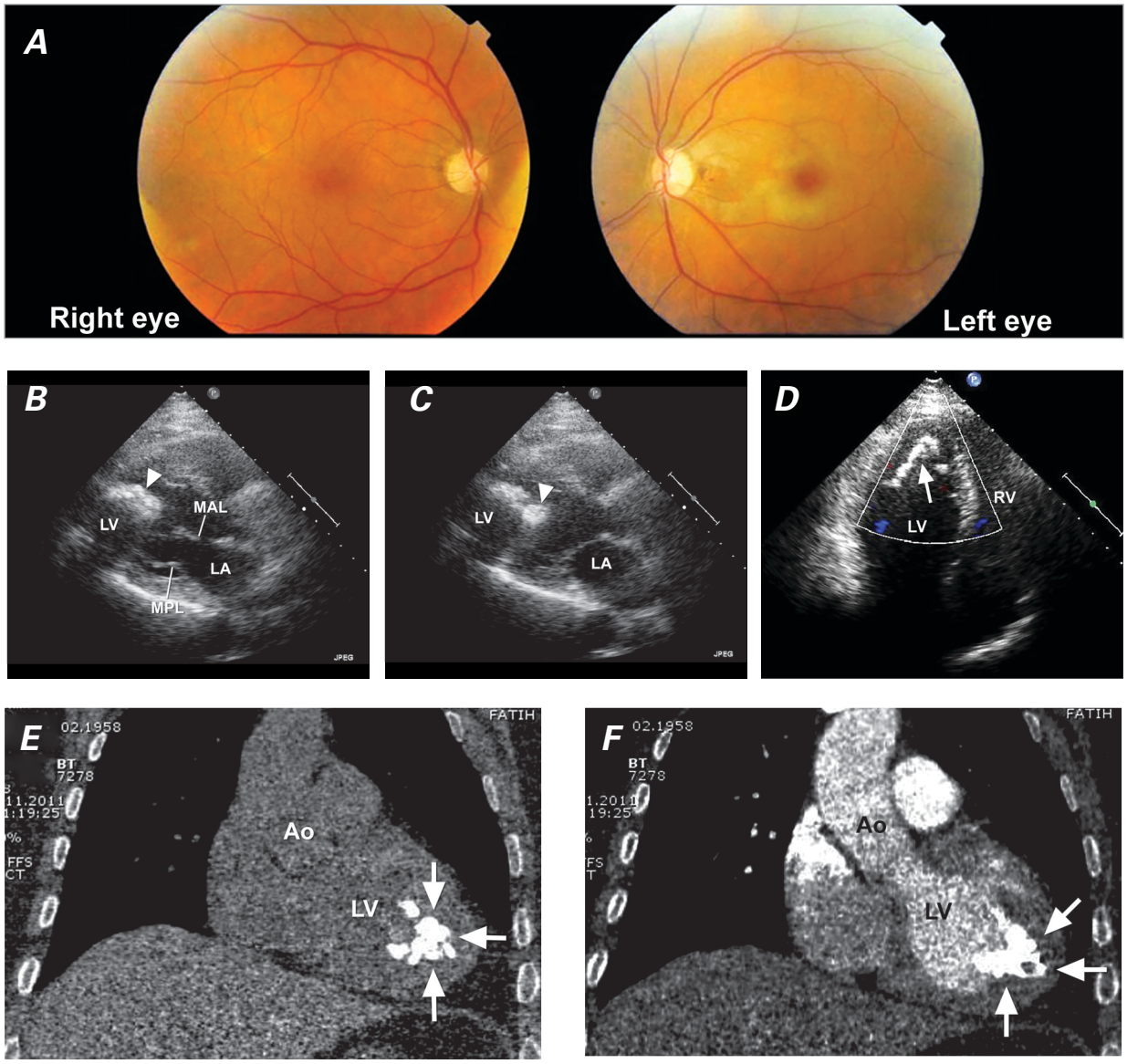

Fig. 1 ***A****) Fundus photography shows a cherry-red spot with retinal pallor typical of central retinal artery occlusion in the left eye and a normal right eye. In the* ***B****) diastolic phase and* ***C****) systolic phase, a transthoracic echocardiogram (parasternal long-axis view) shows a mobile calcified mass (arrowhead) in the left ventricle.* ***D****) A transthoracic echocardiogram (apical 4-chamber view) shows a hyperechoic calcified mass (arrow) originating from the anteroapical and septoapical region of the left ventricle. Computed tomograms (coronal oblique views)* ***E****) with and* ***F****) without contrast material show an echogenic intracavitary irregular mass (arrows) attached to the apex of the left ventricle.*

Ao = aorta; LA = left atrium; LV = left ventricle; MAL = mitral anterior leaflet; MPL = mitral posterior leaflet; RV = right ventricle

remains of the extirpated tumorous mass. Histopathologic examination showed amorphous eosinophilic hyalinized material, along with dense calcification (Fig. 2C). No myxomatous tissue was seen; the final diagnosis was calcified amorphous tumor of the heart.

The patient, who had no perioperative complication and experienced no further embolic events, was discharged from the hospital on the 6th postoperative day. Oral warfarin therapy was started preoperatively and was continued for 3 months postoperatively. One year later, the patient remained asymptomatic.

Discussion

The cardiac CAT comprises calcium deposits in a matrix of amorphous degenerating fibrinous material.[11]

It has been suggested that cardiac CAT has its origin in organized mural thrombus, but the exact cause is unknown. Differential diagnosis of this lesion might include calcified myxomas or fibromas, calcified tuberculoma, thrombi, emboli, vegetations, and tophaceous pseudogout—as well as tumoral calcinosis, especially in patients with end-stage chronic renal failure due to ab-

normalities in calcium, parathyroid hormone, or vitamin D_3 metabolism.[15]

Calcified amorphous tumors have been said to occur in the absence of clinical preconditions for thrombosis or hypercalcemia. However, current laboratory analysis is now able to rule out the infrequent causes of hypercoagulability, such as inherited thrombophilias or antiphospholipid antibody syndrome, apart from the more common causes (smoking, use of contraceptive pills, pregnancy, and malignancies).[10] All of these causes were ruled out in our patient.

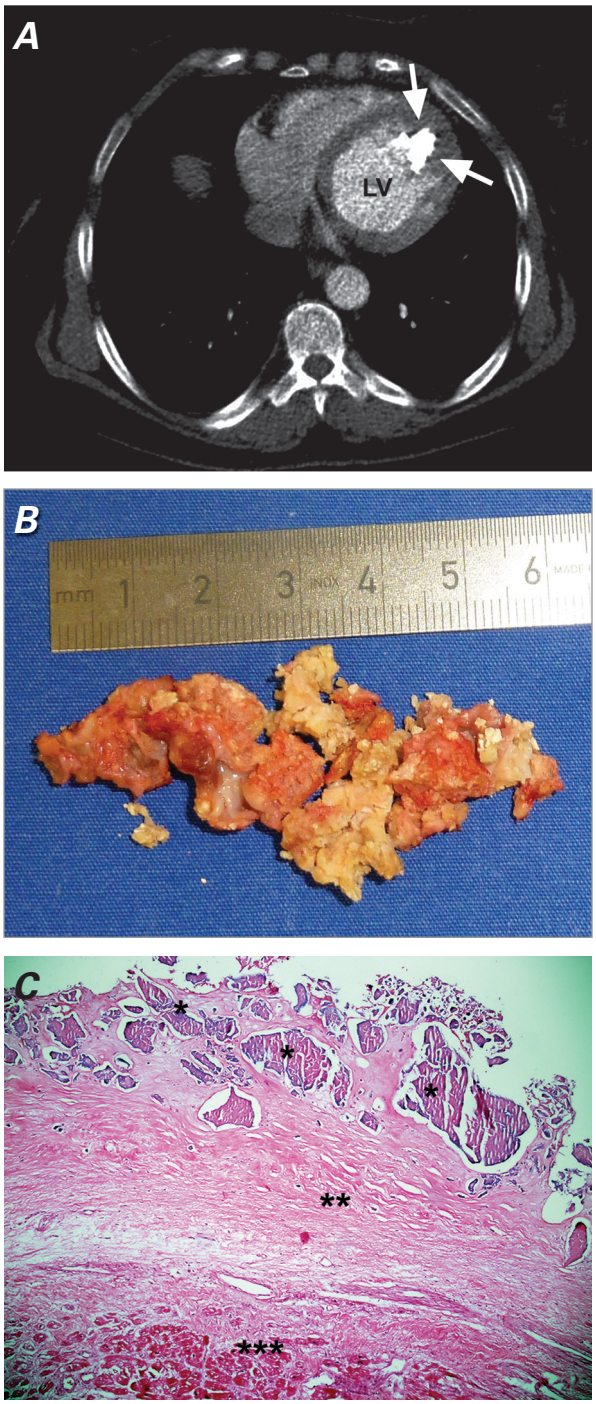

Fig. 2 ***A****) Axial computed tomogram shows an echogenic intracavitary irregular mass (arrows) attached to the apex of the left ventricle (LV).* ***B****) View of the surgically excised calcified amorphous tumor as multiple pieces.* ***C****) Microscopic image of cardiac calcified amorphous tumor reveals amorphous eosinophilic hyalinized material (*) along with dense calcification (**). Myocardial muscle (***) is seen beneath the hyalinized material (H & E, orig. ×100).*

Cardiac CATs can occur in any chamber of the heart. Most cardiac CATs present in ventricular cavities; however, they have also been described in atria, in valves, and in valvular annuli.[20,21] Cardiac CATs cause symptoms due to obstruction or to the embolization of calcific fragments. The clinical presentation, which depends on the location and size of the masses, includes dyspnea, chest pain, syncope, and pulmonary or systemic embolism.[10] The first description of retinal arterial embolism in association with a cardiac CAT was published in 2011.[15] Mobile CATs definitely indicate a greater risk of cerebrovascular accident or systemic embolism than do immobile amorphous tumors.[12] To our knowledge, our case is only the 2nd report of retinal arterial embolism due to cardiac CAT in the English-language medical literature.

In the absence of distinctive clinical and imaging features, preoperative differentiation between neoplastic and non-neoplastic masses remains difficult.[11] It is important to remember that current cardiac imaging techniques still do not specifically recognize cardiac CAT, although they can help to narrow down the differential diagnosis of calcified lesions.[10] Hence, excision of the lesion and histopathologic examination is necessary for accurate diagnosis.[11] Histopathologically, CAT is characterized by nodular calcium deposits over a matrix of fibrin or amorphous fibrin-like material and by hyaline formation and the presence of chronic inflammatory cells and degenerated hematologic elements without malignant cells.[3,8]

Surgical excision is recommended if the lesion is large or symptomatic. In most instances, surgery is curative, especially for pedicled lesions.[10] Most cases reported thus far have run a benign course after surgical excision of the intracardiac mass, although some residual calcified tissue at the location of the original mass might be seen.[1] Cardiac CAT can recur and enlarge after excision, especially if excision has been incomplete.[15] Such patients should be monitored vigilantly with repeat cardiac imaging after excision.[4]

Again, CAT's precise cause and the best approach to treatment remain unclear.

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